THE MECHANISM OF SUDDEN DEATH IN DISSECTING ANEURYSM WITH INTRACARDIAC RUPTURE

BY

ALFRED PICK AND GUISEPPE MININNI*

From the Cardiovascular Department, Medical Research Institute, Michael Reese Hospital, Chicago

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A number of observations on cardiac death have been reported (Stroud and Feil, 1948) in man as well as in the experimental animal. However, as stated recently by P. D. White (1951), " We need more." A case is presented in which a dissecting aneurysm of the aorta caused sudden death by perforation into cardiac structures. Close electrocardiographic observation of the terminal events was possible and revealed some unusual features. While probably related in part to the particular type of death, these observations may provide some insight into the variety of phenomena associated with the termination of cardiac activity and of human life.

Case Report

M.A., a 74-year-old white man, was found at noon in his home in deep coma, cyanotic, with laboured respiration, and a slow pulse. According to the reports of the family he was not previously ill, his only complaints being weakness. On arrival at the hospital he was awake, responded to pain stimuli but not to questions. His colour was normal, the pulse rate 46 a minute, the respiration 30, the blood pressure 140/80 mm. Hg. The heart sounds were slow and regular and variations of the intensity of the first sound were noted. Subsequently the pulse rate changed to 75 regular, the blood pressure to 190/100 and the respiration to 22 a minute.

Physical examination showed normal reaction of the pupils to light, and on fundoscopy, flat discs, congestion of the vessels, and no hæmorrhage nor exudates. Some nuchal rigidity was present. The lungs were normal to percussion and auscultation. The heart appeared of normal size on percussion, its action was regular, and no murmurs were heard. The liver and spleen were not enlarged. The neurological findings were normal except for hyperexcitability of the deep tendon reflexes: no abnormal reflexes could be elicited.

The patient continued to improve for the next two hours. His sensorium became clear but he still answered question in a peculiar fashion. At 8 p.m. he was able to give a detailed history which revealed little more than some abdominal distress during the last months: it was described as being diffuse in nature and not associated with nausea, vomiting, or bowel distress.

During the following day the patient remained well and his condition was unchanged. The temperature varied between 97° and 99° F; respiration between 18 and 20, and heart rate between 68 and 80. The urine had a specific gravity of 1022 and showed some albumin, the sediment showed hyaline and granular casts and occasional red cells. Fasting blood sugar was 119 mg. and blood urea nitrogen 17·6 mg. each per 100 ml.; the Kahn test was negative. Hæmoglobin was 14·6 g., red blood count 4·8 million, white blood count 9200 with a normal differential count. The sedimentation rate was 19 (Wintrobe). For technical reasons no electrocardiograms could be obtained on the first two days. The clinical diagnosis at this time was cerebral arteriosclerosis and cerebral ischæmia due to Stokes-Adams syndrome.

On the third morning the first electrocardiogram was taken. While lead I was being recorded (Fig. 1a) he suddenly stopped breathing and became cyanotic. The pulse became imperceptible and the blood pressure dropped to zero. The heart sounds became slow and irregular and within a few minutes stopped completely. Emergency treatment (consisting of artificial respiration, pressure oxygen, infusion of plasma and adrenaline) was unsuccessful and clinical death took place within a few minutes. However, signs of electrical activity of the heart were recorded for forty more minutes. Pertinent and successive parts of cardiograms obtained during this period are reproduced in Fig. 1 and 2 and their interpretation is given in the respective legends.

* Fullbright Fellow. Now Assistant in Medical Clinic, University of Florence, Florence, Italy.

Necropsy was performed the same day by Dr. O. Saphir and Dr. B. Taylor to whom we are indebted for the following description of anatomical findings and Fig. 3. The pathological diagnosis was idiopathic cystic necrosis of the media of the aorta; dissecting aneurysm of the ascending aorta with extension to the arch, the innominate, right and left common carotid arteries and to the right atrium; perforation into the pericardium with hæmopericardium; generalized arteriosclerosis; acute passive hyperæmia of the liver, spleen and kidneys; anomalous origin of the left posterior cerebral artery; arterio- and arteriolo-nephrosclerosis.

The significant findings were localized to the heart and the great vessels. The pericardial cavity was distended by a mass of freshly clotted blood which enveloped the heart in a layer of 2·5 cm. thickness. It measured about 750 ml. in volume and was easily removed leaving a glistening serous pericardium. The source of the bleeding into the pericardium was localized as an irregular 2 cm. long rent over the right lateral aspect of the base of the aorta at its emergence from the left ventricle. The tear opened into the transverse sinus of the pericardium and was surrounded by a 3 cm. wide subserosal ecchymosis.

The heart weighed 410 g., being moderately hypertrophied primarily due to thickening of the left ventricular myocardium which measured up to 17 mm. in width. The endocardium was smooth and transparent throughout and the circumference of the valvular orifices well within normal limits. The cusps were thin and pliable, showing no evidence of inflammatory or degenerative distortion.

The aortic intima exhibited moderate atheromatosis but there was no calcification or ulceration. On the posterior aspect of the aortic arch, 1·5 cm. above the aortic ring, there was an irregular sinuous tear of the intima which ran obliquely upwards for a distance of 3 cm. and extended outwards into the intima. The edges of the rent were separated by a fresh blood clot which led into a medial dissection. This completely encircled the aorta thickening the wall to 3 or 4 cm. Proximally, it extended into the root of the aorta surrounding the orifice of the coronary artery, but neither ostia nor lumina of the coronary arteries were occluded. In addition, the hæmorrhage could be traced into the right atrium where it formed an extensive subendocardial hæmorrhage of the right surface of the inter-auricular septum. Measuring 4·5 cm. in diameter, this hæmatoma reached the medial leaflet of the tricuspid valve, thus involving the region of the A-V node (Fig. 3). Distally the medial dissection progressed to the origin of the left common carotid artery continuing into the media of the innominate and right and left carotid arteries for a distance of about 4 to 5 cm. On transection of the vessels, however, their lumina were not appreciably narrowed.

Microscopic sections of the aorta distal to the dissection contained foci of mucinous degeneration consisting of a deposition of interstitial basophilic material. In areas this coalesced to form irregular cystic spaces of various sizes filled with myxomatous material. Sections through the hæmorrhage localized it to the middle and outer thirds of the media. There was neither an exudative nor proliferative response. The intimal changes were of the usual atheromatous nature consisting of fibrous thickening in which were found cholesterol slits and lipid laden histiocytes.

COMMENT

A correlation of clinical, anatomical, and electrocardiographic data suggests the following reconstructions of events leading to sudden death in this patient. Dissection of the aortic wall, which was the seat of a typical idiopathic medial necrosis probably took place in two separate episodes. At first, dissection beginning at the site of the intimal tear near the aortic root was carried in a distal direction and produced the comatous state for which the patient was admitted to the hospital. Cerebral symptoms like disorientation, syncope, and coma are known to occur as initial signs of aortic dissection (3); in association with bradycardia this has been ascribed to involvement of the endings of the depressor nerves (4). Since in this instance, hæmorrhage extended into the walls of both carotid arteries, a resulting cerebral ischæmia (5) might also account for the initial symptoms. The dissection then apparently stopped at this level and the patient recovered. However, 36 hours later when the patient was moved in order to obtain a cardiogram, a second and fatal dissection took place, this time directed towards the root of the aorta. The sequence of observed cardiographic events (Fig. 1 and 2) suggests that hæmorrhage into the atrial septum shortly preceded the terminal perforation into the pericardial cavity.

While rupture into the pericardium and death from cardiac tamponade is seen commonly subsequent to extension of an aortic dissection in a proximal direction (5–8) only a few instances are on record where hæmorrhage took place into cardiac structures surrounding the aortic root (5, 9–11). In two of the reported cases (5, 11) the post-mortem findings were similar to those in our case in that a resulting hæmatoma of the atrial septum closely approached the region of the A-V node (Fig. 3). These patients also had marked bradycardia at the time of the dissection and,

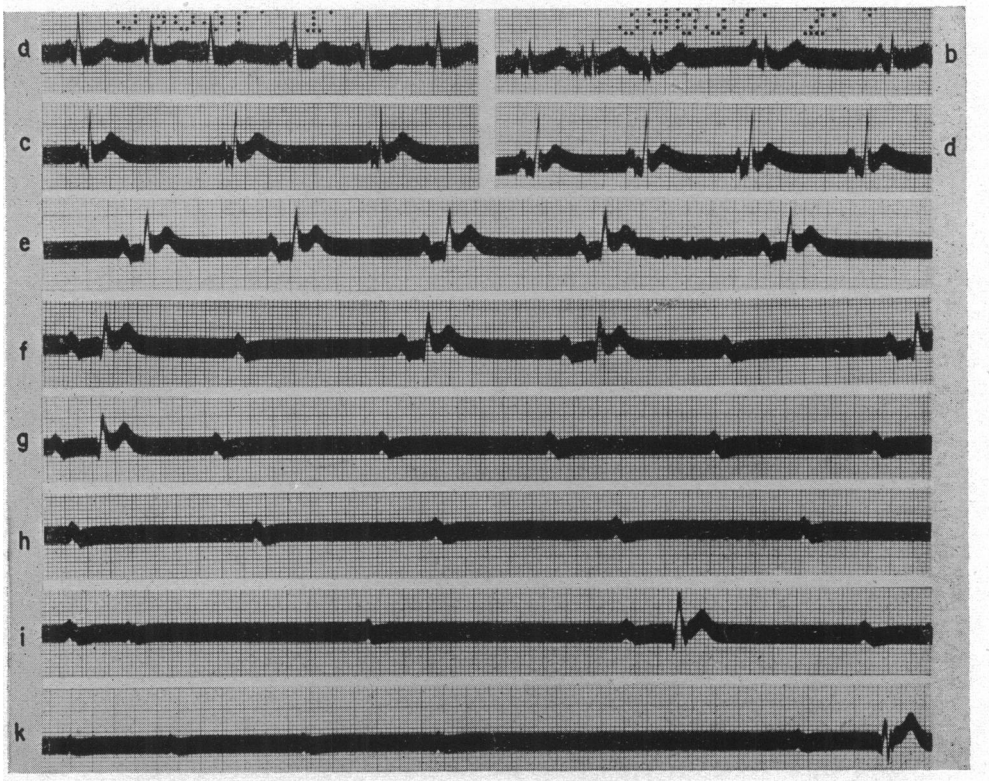

FIG. 1.—Part of successive cardiograms recorded during and following clinical death. (Strip a is lead I, all others, lead II.) The terminal stages of auricular activity and of A-V conduction are seen.

(a) *Alterations of auricular conduction.* Auricular rate regular at 96 a minute, but alterations in contour of P. P–R interval between 0·12 and 0·14 sec. The third ventricular complex is premature and originates in the A-V node.

(b) *Depression of auricular conduction and S-A block.* Auricular rate changes abruptly from 103 to 52, P varies in size and contour, P–R interval unchanged (0·14 sec.).

(c) *Ectopic auricular pacemaker.* Rate regular and slow (48), P bizarre in contour, P–R interval 0·10 sec. Increased size of QRS and elevation and shortening of ST–T.

(d) and (e) *Periods of auricular irritation with runs of auricular tachycardia.* In (d) the auricular rate is faster (68), P remains notched and diphasic, P–R 0·16 sec. Occasionally P waves more bizarre and P–R interval shortened, as in preceding strip. In (e) auricular rate slowed to 46, P diphasic and widened, and P–R interval slightly prolonged (0·20 to 0·24 sec.) A short run of four premature auricular complexes at an irregular rate of about 206, differing in contour from other P waves in this strip, is seen following the fourth ventricular complex; none of these are conducted to the ventricles.

(f) *Development of A-V block.* Large and diphasic P waves occur at a slow (48) and uneven rate. A-V block with Wenckebach phenomenon at a 3:2 ratio, P–R increasing from 0·24 to 0·32 sec. As in strip (e), the S–T segment is elevated indicating alteration of ventricular deactivation.

(g) to (k) *Progression to complete A-V block with transient depression of primary and secondary pacemakers.* Auricular rate slows further to 37 and ventricular complexes disappear (g to h). A-V conduction after (h) completely interrupted. Occasional idioventricular beats in (i) and (k), P waves irregular. The diminishing rate and size of P and alterations in contour indicate progressive deterioration of formation and spread of auricular impulses.

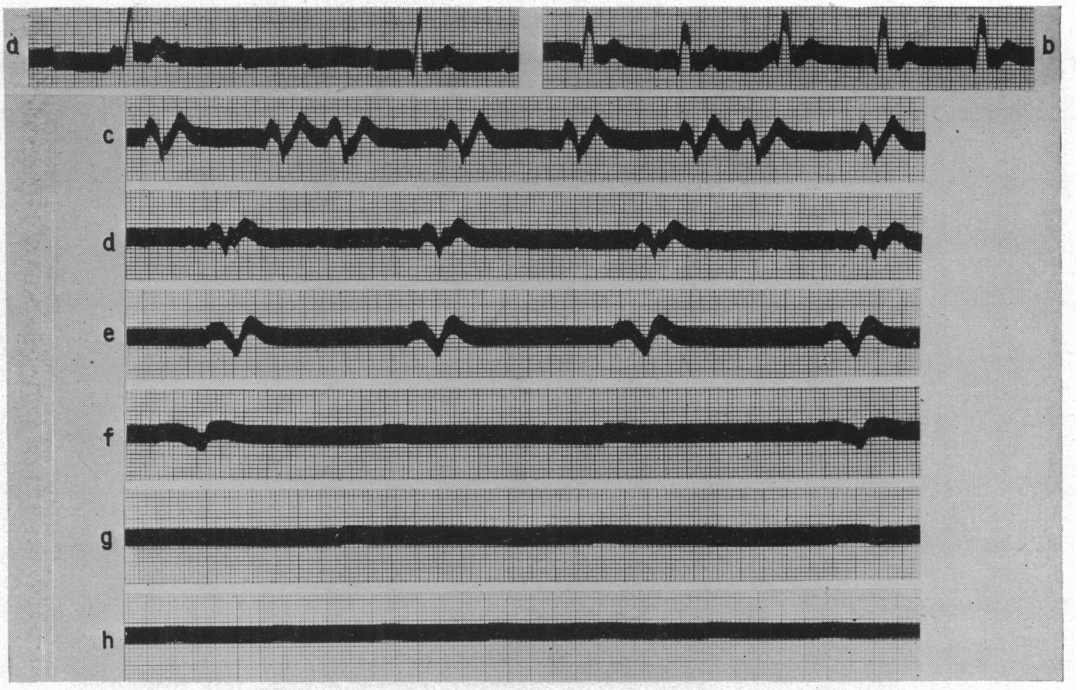

FIG. 2.—Strips (a) to (h) are the continuation of Fig. 1 and portions of successive longer tracings, recorded in lead II. The death of the auricles and ventricles is revealed in these tracings.

(a) to (d) *Disappearance of auricular activity, irregular idioventricular pacemaker, development of intraventricular* block. The last manifestations of auricular activity occur intermittently (a, b, and d) at a relatively rapid rate of 103, Before disappearing, P becomes more and more shallow. There is complete A-V block, with an idioventricular pacemaker at varying rates. Although in (a) and (b) the QRS complex widens progressively (0·12 to 0·16), its contour remains similar to conducted beats in Fig. 1. Thus, the impulse seems to originate somewhere above the bifurcation of the common bundle but is transmitted to the ventricles with progressive impairment of intraventricular conduction. The long ventricular interval in (a) is a multiple of the short ones seen in (b); hence, the derangement of intraventricular conduction appears to result, at times, in complete " exit block " of the pacemaker. (Compare with similar events in the auricles seen in Fig. 1b.) Following an allorhythmic phase with intermittent bigeminal rhythm (c), the ventricular action becomes slow and almost regular, but impairment of intraventricular conduction progresses further. Thus, in (c) and (d), the QRS complex is lower, widened to 0·24 sec., and consists of two distinctly separated diphasic portions. T waves upright as before.

(e) to (h) *Progression of intraventricular block to dissociation of ventricular activation and ventricular arrest.* No sign of auricular activity. Ventricular rate further slowed to 32. Separation of ventricular activation into two portions now more marked, resulting in two negative initial deflections, each of which merges into its own positive final deflection. In (e), the interval from the beginning of the first to the end of the second QRS portion measures 0·36 sec. Ventricular complexes of the same type, but lower in amplitude and even longer in duration (0·42 sec.) are seen at the beginning and at the end of (f); in its middle part only the initial portion with its T wave occurs at the expected time. Thus, at this stage, intraventricular conduction seems so deranged that the idioventricular impulse activates one part of the ventricles but temporarily fails to reach the other. This ventricular block becomes complete in (g), where only low and regular deflections are seen corresponding to the initial parts of ventricular complexes of the preceding strips. Finally, in (h), only traces of activity of one part of the ventricle represent the last recorded electrical events.

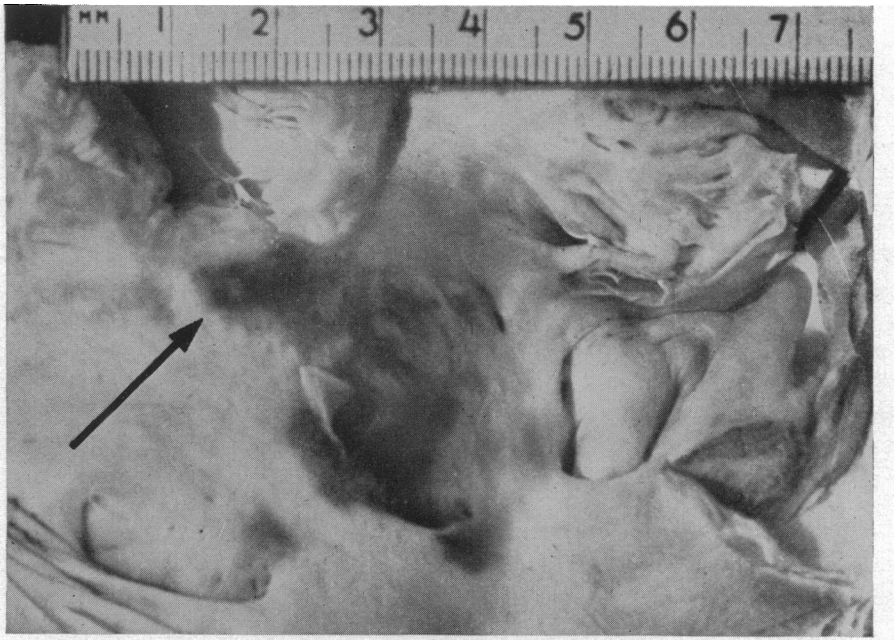

FIG. 3.—View into the cavity of the right atrium revealing the extensive subendocardial hæmorrhage in the atrial septum, extending to the region of the A-V node (at arrow).

although no electrocardiograms were obtained, this was related to the autopsy findings by one of the authors (11).

While respiration ceased immediately and clinical death took place within a few minutes, signs of electrical activity of the heart continued for forty minutes. Electrocardiograms have been recorded for much longer periods after cessation of cardiac contractions (12). Cardiographic death was then seen to occur in a certain order and in three main stages (13–16). An initial phase of sinus slowing may alternate with rapid phases of auricular activation; in a following phase, auricular activity ceases and ventricular activation is effected by a subsidiary pacemaker; finally there is ventricular arrest which may or may not be preceded by a period of ventricular fibrillation. The question of incidence of terminal ventricular fibrillation has been investigated both experimentally and clinically. Harris (17) who studied cardiac death under various controlled experimental conditions saw fibrillation frequently in conditions favourable for the production of ectopic impulses, e.g. localized ischæmia following occlusion of a coronary artery, while in conditions leading to generalized anoxia and tissue death ventricular arrest was effected by failure of pacemakers or of A-V conduction. In the dying human heart ventricular fibrillation was observed as frequently as simple standstill (1) regardless of the presence or absence of heart disease causing death. To our knowledge no clinical or experimental electrocardiographic observation has been reported describing the mechanism of sudden death from acute cardiac tamponade which undoubtedly was the cause of death in the presented case. Usually, in this condition, death is not instantaneous (18).

Although in general following the pattern outlined above, the electrocardiographic evolution here appeared to be modified, at least initially, and could be related to the particular findings post mortem. Blockage of impulse transmission, first within the auricles and later at the A-V junction appeared to be dominating and was mainly responsible for the slowing, and the initial irregularities of the cardiac action (Fig. 1). A-V block has been considered by some (12) to be rare among the phenomena of cardiac death, but P–R prolongation as well as S-A block has been noted by others (15, 16, 20–22). However, none of their illustrations shows systematic progression to the highest degrees of block such as are seen in Fig. 1. Dissociation of auricular and ventricular action appeared to result from extreme slowing of the primary impulse formation under vagal influences

associated with death (23), rather than from block of A-V conduction. Thus, in all likelihood, the organic lesion in the atrial septum developing in the course of aortic dissection was, by extending to the region of the conduction system, responsible for the cardiographic alterations in the first part of this observation.

As a result of complete interruption of A-V conduction, a period of complete ventricular standstill is seen in Fig. 1h followed by the appearance of an idioventricular pacemaker of supraventricular origin, discharging initially at an extremely slow and irregular rate. Signs of severe depression of ventricular pacemakers varying with those of irritation of these pacemakers are common phenomena associated with cardiac death (16). How far the administration of stimulating drugs like adrenaline, during the last efforts to restore life, might be responsible for irregularities of the terminal cardiac activity in this and in previously reported cases is impossible to assert. However, if the interpretation of the tracings taken during the second part of observation (Fig. 2) is correct, the profound ventricular arrhythmias were in this instance not so much the result of terminal disturbance of impulse formation of the cardiac impulse but of disturbances in their propagation through the ventricles. First signalled by prolongation of QRS duration (Fig. 2b) the disturbance of intraventricular conduction is seen to develop further in several stages. A temporary block surrounding the pacemaker is suggested by the phenomenon of " exit block " in Fig. 2a. Later, while impulse formation continues at a slow but fairly regular rate, progressive slowing of its propagation through the ventricles can be followed throughout the tracings (Fig. 2d–e) resulting in separation of activation and deactivation of the ventricles into two distinct portions; then the impulse apparently failed, first temporarily and eventually permanently (Fig. 2f–g), to activate a major part of the ventricles, before these last manifestations of electrical activity also disappeared.

Ever since investigations were made into the mysteries of cardiac death interest has centered mainly around two points: What is the last part of the heart to die, the ultimum moriens? And what are the last signs and mechanisms of its activity? Shallow terminal waves of the type seen in Fig. 2g–h may be interpreted as representing terminal auricular activity unless their development can be followed in long successive strips. The analysis of the tracings in Fig. 2 leaves little doubt that, in accordance with the majority of observations (13, 14, 16, 21, 23–26) and in contrast to some others (22, 27–29), the last part of the heart to cease to function was the ventricles.

These last moments of ventricular activity have been described as giving rise to characteristic " agonal ventricular complexes " (30) consisting of large monophasic potentials of prolonged duration with elevation of S–T and fusion of QRS and T. The mechanism responsible for this bizarre contour has never been established. Whatever the immediate cause may be (whether injury currents due to terminal hypoxia, alterations of intracellular or extracellular potassium in the myocardium as recently suggested (31), or other metabolic disorders so far undetected), the tracings that form the basis of this report suggest that one of the operating mechanisms may be a profound disturbance of cardiac conductivity. Whether this is related to the acute changes of cardiodynamics and the circulatory failure effected by the sudden cardiac tamponade (32, 33) remains a matter of speculation that warrants further study.

Summary

In a case of dissecting aneurysm of the aorta extension to the aortic root resulted in hæmorrhage into the atrial septum involving the region of the A-V node, and eventually lead to a fatal rupture into the pericardium and sudden death.

Electrical activity of the heart persisted and was recorded for forty minutes after clinical death. A detailed analysis of the records suggested that the dominant factor in the production of terminal irregularities of the heart action was a profound and progressive disturbance of cardiac conductivity involving successively the auricles, A-V conduction, and intraventricular conduction. Cardiac arrest occurred following a period of dissociation of ventricular activation due to the development of ventricular block. A possible relationship of the unusual anatomical and electrocardiographic findings is considered.

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